# Surgical repair of left internal jugular phlebectasia

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**Jugular vein phlebectasia, a fusiform dilatation of a vein without tortuosity, is a rare cause of cervical neck swelling in children. It commonly presents as a soft cystic mass in the neck that transiently appears during straining. Because of its rarity, jugular vein phlebectasia cases have frequently been misdiagnosed or have been managed inappropriately. This report describes the case of a left-sided internal jugular phlebectasia in a 4-year-old child that was surgically treated with a resection and an end-to-end repair. (J Vasc Surg 2008;47:1337-8.)**

A 4-year-old healthy girl presented with a left cervical swelling that her parents first noticed when she was 18 months old. Since then, the swelling had increased in size and now the mass enlarged to a noticeable size upon minimal exertion. There were no associated systemic symptoms, and the patient's history was unremarkable, with no history of surgery, infection, or trauma to the neck.

On examination, no swelling was observed at rest. On Valsalva maneuver, the mass appeared as a left-sided, soft, painless, nonpulsatile, and compressible neck mass. It was 2 cm in the transverse direction and 3 cm along the longitudinal axis. The mass was anteromedial to the left sternocleidomastoid muscle. There were no bruits or palpable lymphadenopathy. The result of physical examination was otherwise unremarkable.

The larynx was evaluated by fiberscopic laryngoscopy and was found to be normal. A chest radiograph was interpreted as normal, with no abnormalities of the lung fields, cardiac silhouette, or mediastinum. An ultrasound examination of the neck revealed a very large left internal jugular (IJV) vein with marked increase in dilatation just above the jugular–subclavian junction during a Valsalva maneuver. There was no evidence of thrombus in the lumen of the IJV. The direction of the blood flow within the phlebectasia was caudally oriented. A contrast-enhanced computed tomography (CT) scan of the neck demonstrated a fusiform dilatation of the IJV, and the other anatomic structures were normal (Fig 1).

The mass continued to enlarge, and several episodes of exercise-induced dilatation caused the distressed parents to bring the patient to the emergency department. The decision was made to pursue surgical intervention, which was initiated with an oblique incision over the anterior margin of the left sternocleidomastoid muscle. The fusiform dilation of the thin-walled vein was immediately visible. The left IJV was then dissected from above the level of the hyoid cartilage down to the left subclavian vein. A clearly dilated portion was identified from the superior aspect of the thyroid cartilage to approximately 1 cm above the subclavian vein. After systemic heparinization, this area was controlled between vascular clamps and a total length of 3.5 cm of jugular vein, including the dilated portion, was resected and a primary anastomosis was performed (Fig 2). The patient had an uneventful postoperative recovery.

On macroscopic examination, the mass was a dilated, sac-like 3.0- × 1.7-cm tube that was microscopically lined by endothelial cells. The subendothelial layer was variable, with incomplete layers of smooth muscle and large segments of fibrosis replacing the normal vessel wall constituents.

Postoperative ultrasound studies showed a successful repair of the previously identified saccular ectasia of the left IJV. There was no evidence of intraluminal thrombus or significant focal stenosis. A follow-up ultrasound study performed 6 weeks postoperatively showed appropriate flow and normal caliber of the left IJV. At 6 months, the patient remains healthy, with no recurrence.

## DISCUSSION

A fusiform dilatation of the jugular vein is called a *phlebectasia* (JVP).[1] Although JVPs are rare, they are becoming increasingly recognized, partly due to improved diagnostic techniques.[1] The cause of JVPs is still obscure but is usually attributed to idiopathic origins or congenital defects in the muscular wall of the IJV.[2,3] Generally, histopathologic studies show normal dilated veins.[4] In some cases, there is a loss of or disordered arrangement in the smooth muscle cells, elastic fibers, and connective tissue.[5,6]

Jugular vein phlebectasias commonly present in childhood as a localized swelling in the anterior neck. These masses are nontender and compressible, appearing during situations in which there is an increase in intrathoracic pressure.[5] The bulging mass is demarcated clearly, without bruit, and does not move upward or downward with deglutition. Although most JVPs are asymptomatic, some children have complained of slight discomfort during deglutition, hoarseness in phonation, and a feeling of a cervical foreign body.[2]

A JVP can involve the internal, external, or anterior jugular veins, with a predilection for the IJVs, particularly on the right side.[3,7] The differential diagnosis of a neck mass in pediatric patients is broad, but a swelling that occurs *only* on straining reduces the possibilities to phlebectasias, laryngoceles, cysts of the superior mediastinum, and inflation of pulmonary apical bullae.[1,2] Of these, the

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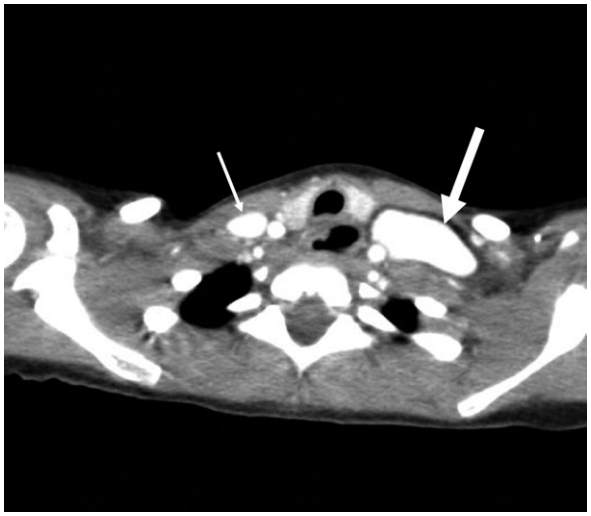

**Fig 1.** A computed tomography scan of the left jugular vein at the apex of the lung during a Valsalva maneuver reveals the dilated left jugular vein (*thick arrow*) and the normal right jugular vein *(narrow arrow)*.

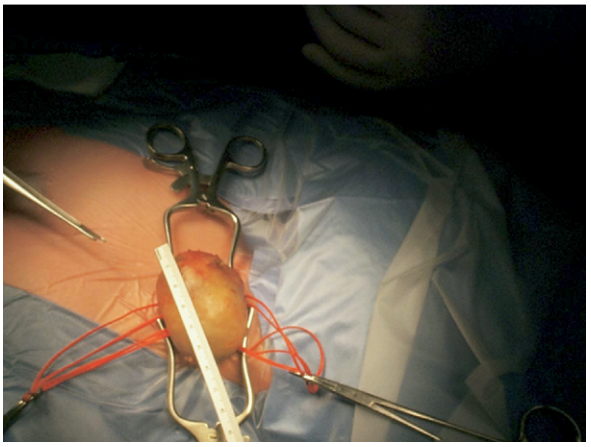

**Fig 2.** Perioperative photo shows the left internal jugular vein phlebectasia.

diagnosis of laryngocele is the most common. Laryngoscopy directly rules out the possibility of a laryngocele, and a thoracic CT scan rules out the possibility of a mediastinal cyst or tumor.[8]

Ultrasonography during a Valsalva maneuver easily establishes the diagnosis of JVP, and this should be used as a first-line imaging test. During a Valsalva maneuver, the diameter of the affected vein usually increases 1 to 2.2 times compared with its measurements at rest.[3,4] The JVP mass reduces to normal on supination or local compression. Color Doppler imaging confirms the presence or absence of blood flow and thrombus formation in the lumen of the IJV.

Because JVP is a benign condition with no medical treatment, most authors advocate conservative observation unless the lesions are symptomatic.[1,4] For lesions that are asymptomatic, surgical intervention is recommended if cosmetic or psychologic concerns are present.[1,3]

In most surgical cases, the JVP and associated vein have been ligated, with the loss of the normal venous drainage pattern on that side. Other interventions described include longitudinal constriction suture venoplasty and partial resection of the phlebectasia.[4,9] Both options have been reported to be safe and successful in eliminating the phlebectasia.[4,9] Our case offers another surgical approach to the treatment of the JVP, with excellent results. To determine the best surgical procedure among these various options, further studies will have to be conducted with longer-term follow-up; however, in instances when the vein may be mobilized extensively and proximal and distal vascular control can be obtained, resection with primary reanastomosis would appear to offer a complete removal of the abnormal tissue with no loss of normal venous drainage patterns or a potential site of vascular access.

## CONCLUSION

In our case report, a patient with a left internal JVP, which was becoming increasing symptomatic, was successfully treated with an end-to-end anastomosis. This procedure was last described in the literature more than a decade ago. Because of the ease of this operation, this technique should be considered as a surgical option.

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